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* * * * * Welcome to STN International * * * * *

NEWS	1		Web Page URLs for STN Seminar Schedule - N. America
NEWS	2		"Ask CAS" for self-help around the clock
NEWS	3	SEP 09	CA/CAPLUS records now contain indexing from 1907 to the present
NEWS	4	DEC 08	INPADOC: Legal Status data reloaded
NEWS	5	SEP 29	DISSABS now available on STN
NEWS	6	OCT 10	PCTFULL: Two new display fields added
NEWS	7	OCT 21	BIOSIS file reloaded and enhanced
NEWS	8	OCT 28	BIOSIS file segment of TOXCENTER reloaded and enhanced
NEWS	9	NOV 24	MSDS-CCOHS file reloaded
NEWS	10	DEC 08	CABA reloaded with left truncation
NEWS	11	DEC 08	IMS file names changed
NEWS	12	DEC 09	Experimental property data collected by CAS now available in REGISTRY
NEWS	13	DEC 09	STN Entry Date available for display in REGISTRY and CA/CAPLUS
NEWS	14	DEC 17	DGENE: Two new display fields added
NEWS	15	DEC 18	BIOTECHNO no longer updated
NEWS	16	DEC 19	CROPU no longer updated; subscriber discount no longer available
NEWS	17	DEC 22	Additional INPI reactions and pre-1907 documents added to CAS databases
NEWS	18	DEC 22	IFIPAT/IFIUDB/IFICDB reloaded with new data and search fields
NEWS	19	DEC 22	ABI-INFORM now available on STN
NEWS	20	JAN 27	Source of Registration (SR) information in REGISTRY updated and searchable
NEWS	21	JAN 27	A new search aid, the Company Name Thesaurus, available in CA/CAPLUS
NEWS	22	FEB 05	German (DE) application and patent publication number format changes
NEWS	23	MAR 03	MEDLINE and LMEDLINE reloaded
NEWS	24	MAR 03	MEDLINE file segment of TOXCENTER reloaded
NEWS	25	MAR 03	FRANCEPAT now available on STN
NEWS EXPRESS	MARCH 5 CURRENT WINDOWS VERSION IS V7.00A, CURRENT MACINTOSH VERSION IS V6.0b(ENG) AND V6.0Jb(JP), AND CURRENT DISCOVER FILE IS DATED 3 MARCH 2004		
NEWS HOURS	STN Operating Hours Plus Help Desk Availability		
NEWS INTER	General Internet Information		
NEWS LOGIN	Welcome Banner and News Items		
NEWS PHONE	Direct Dial and Telecommunication Network Access to STN		
NEWS WWW	CAS World Wide Web Site (general information)		

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* * * * * STN Columbus * * * * *

FILE 'HOME' ENTERED AT 19:17:32 ON 10 MAR 2004

=> file biosis

COST IN U.S. DOLLARS

SINCE FILE

TOTAL

ENTRY

SESSION

FULL ESTIMATED COST

0.21

0.21

FILE 'BIOSIS' ENTERED AT 19:17:55 ON 10 MAR 2004

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FILE COVERS 1969 TO DATE.

CAS REGISTRY NUMBERS AND CHEMICAL NAMES (CNs) PRESENT
FROM JANUARY 1969 TO DATE.

RECORDS LAST ADDED: 3 March 2004 (20040303/ED)

FILE RELOADED: 19 October 2003.

=> s pompe

312 POMPE

159 POMPES

L1

422 POMPE

(POMPE OR POMPES)

=> s l1 and (hexose (w) tetrasaccharide)

7736 HEXOSE

2001 HEXOSES

9163 HEXOSE

(HEXOSE OR HEXOSES)

1496 TETRASACCHARIDE

396 TETRASACCHARIDES

1773 TETRASACCHARIDE

(TETRASACCHARIDE OR TETRASACCHARIDES)

0 HEXOSE (W) TETRASACCHARIDE

L2

0 L1 AND (HEXOSE (W) TETRASACCHARIDE)

=> s l1 and (tetrasaccharide)

1496 TETRASACCHARIDE

396 TETRASACCHARIDES

1773 TETRASACCHARIDE

(TETRASACCHARIDE OR TETRASACCHARIDES)

L3

5 L1 AND (TETRASACCHARIDE)

=> d l3 kwic 1-5

L3 ANSWER 1 OF 5 BIOSIS COPYRIGHT 2004 BIOLOGICAL ABSTRACTS INC. on STN

TI Analysis of a glucose **tetrasaccharide** elevated in **Pompe**
disease by stable isotope dilution-electrospray ionization tandem mass
spectrometry.

AB Patients with glycogen storage disease type II (GSD II) typically excrete
increased amounts of a glycogen-derived glucose **tetrasaccharide**,
Glcalpha1-6Glcalpha1-4Glcalpha1-4Glc (Glc4), in the urine. With the
advent of a new enzyme replacement therapy for GSD II, there is a . . .

IT

Metabolism; Methods and Techniques

IT

Parts, Structures, & Systems of Organisms

plasma: blood and lymphatics; urine: excretory system

IT

Diseases

Pompe disease: genetic disease, metabolic disease

Glycogen Storage Disease Type II (MeSH)

IT

Diseases

glycogen storage disease type II: genetic disease, metabolic disease, therapy, GSD II
 Glycogen Storage Disease Type II (MeSH)

IT Chemicals & Biochemicals
 butyl 4-aminobenzoate derivatives; glucose; glucose
tetrasaccharide: analysis; recombinant alpha-amylase

L3 ANSWER 2 OF 5 BIOSIS COPYRIGHT 2004 BIOLOGICAL ABSTRACTS INC. on STN
 TI Determination of oligosaccharides in **Pompe** disease by electrospray ionization tandem mass spectrometry.

AB. . . need for biochemical markers to monitor the efficacy of therapy and methods to quantify these markers in biologic samples. In **Pompe** disease, the concentration of a **tetrasaccharide**, consisting of four glucose residues, is reputedly increased in urine and plasma, but faster and more sensitive methods are required. . . spectrometry, of oligosaccharide concentrations in urine (n = 6), plasma (n = 11), and dried-blood spots (n = 17) from **Pompe**-affected individuals. Age-matched control samples of urine (n = 10), plasma (n = 28), and blood spots (n = 369) were also analyzed. Results: The mean **tetrasaccharide** concentration was increased in urine from infantile-onset (0.69-12 mmol/mol of creatinine) and adult-onset (0.22-3.0 mmol/mol of creatinine) **Pompe** individuals compared with age-matched controls. In plasma samples, an increased **tetrasaccharide** concentration was observed in some infantile patients (up to 22 mumol/L) compared with age-matched controls (mean, 2.2 mumol/L). The method. . . determine oligosaccharide concentrations in a single 3-mm blood spot, but no differences were observed between blood spots from control and **Pompe**-affected individuals. Conclusions: Measurements of oligosaccharide concentrations in urine by this new method have potential application for the diagnosis and monitoring of patients with **Pompe** disease. Plasma analysis may have limited application for infantile patients, but analysis of blood spots does not discriminate between controls. . .

IT . . .
 IT Parts, Structures, & Systems of Organisms
 blood: blood and lymphatics; plasma: blood and lymphatics; urine: excretory system

IT Diseases
Pompe disease: genetic disease, metabolic disease
 Glycogen Storage Disease Type II (MeSH)

IT Diseases
 lysosomal storage disorder: metabolic disease

IT Chemicals & Biochemicals
 1-phenyl-3-methyl-5-pyrazolone; creatinine; oligosaccharides: determination; **tetrasaccharide**

L3 ANSWER 3 OF 5 BIOSIS COPYRIGHT 2004 BIOLOGICAL ABSTRACTS INC. on STN
 TI Validation of glucose **tetrasaccharide** as a biomarker for diagnosis and monitoring enzyme replacement therapy for **Pompe** disease.

IT Major Concepts
 Genetics

IT Parts, Structures, & Systems of Organisms
 blood: blood and lymphatics; urine: excretory system

IT Diseases
Pompe disease: genetic disease, metabolic disease
 Glycogen Storage Disease Type II (MeSH)

IT Chemicals & Biochemicals
 acid a-glucosidase; glucose **tetrasaccharide**: biomarker

L3 ANSWER 4 OF 5 BIOSIS COPYRIGHT 2004 BIOLOGICAL ABSTRACTS INC. on STN
 TI Liquid chromatographic assay for a glucose **tetrasaccharide**, a putative biomarker for the diagnosis of **Pompe** disease.

AB. . . tetraglucose, normally excreted in the urine, has previously been shown to be elevated in a number of pathological conditions including

Pompe disease (glycogen storage disease type II), which is caused by a deficiency of the lysosomal enzyme acid alpha-glucosidase. Concentrations of. . . 1-5, 6-10, 11-20, and >20 years, both in normal individuals and in a cohort of 21 patients with enzymatically confirmed **Pompe disease**. The Glc4 concentration decreased with age in both groups, but all the patients had elevated Glc4 levels compared with. . . urine. Our results demonstrate that this method is suitable for application in clinical laboratories to help establish the diagnosis of **Pompe disease**.

IT Major Concepts

Clinical Chemistry (Allied Medical Sciences); Methods and Techniques

IT Diseases

Pompe disease: genetic disease, metabolic disease

Glycogen Storage Disease Type II (MeSH)

IT Chemicals & Biochemicals

glucose **tetrasaccharide**: assay, biomarker

L3 ANSWER 5 OF 5 BIOSIS COPYRIGHT 2004 BIOLOGICAL ABSTRACTS INC. on STN

AB. . . characterization and some properties of leucocyte alpha-glucosidase towards disaccharides with the alpha-1, 4 (maltose) and alpha-1, 6-glucosidic linkage (isomaltose) and **tetrasaccharides** with the alpha-1, 4 (maltotetraose) and alpha-1,6-i glucosidic linkage (**tetrasaccharide**, Glc4alpha1 -> 6Glc4alpha1 -> 4Glc4alpha1 -> 4Glc, which was isolated from the urine of a patient with glycogenosis type II).. . . neutral pH. Acid alpha-glucosidase could hydrolyze maltose about 10 times faster than isomaltose, and maltotetraose about 5 times faster than **tetrasaccharide** isolated from urine. In leucocytes of the patient with late onset glycogenosis type II, acid alpha-glucosidase activities towards maltose, isomaltose, maltotetraose and **tetrasaccharide** isolated from urine showed 75.3%, 67.4%, 76.5% and 41.4% of normal control values, respectively. Neutral alpha-glucosidase activities towards these four oligosaccharides were normal. **Tetrasaccharide** with alpha-1, 6-glucosidic linkage might be accumulated by the impaired hydrolysis in the circulation as well as the leakage of. . .

IT Miscellaneous Descriptors

HUMAN MUSCLE **POMPE'S DISEASE**

=> d l3 iall 1-5

L3 ANSWER 1 OF 5 BIOSIS COPYRIGHT 2004 BIOLOGICAL ABSTRACTS INC. on STN

ACCESSION NUMBER: 2003:298793 BIOSIS

DOCUMENT NUMBER: PREV200300298793

TITLE: Analysis of a glucose **tetrasaccharide** elevated in **Pompe disease** by stable isotope dilution-electrospray ionization tandem mass spectrometry.

AUTHOR(S): Young, Sarah P. [Reprint Author]; Stevens, Robert D.; An, Yan; Chen, Yuan-Tsong; Millington, David S.

CORPORATE SOURCE: Biochemical Genetics Laboratory, Division of Medical Genetics, Department of Pediatrics, Duke University Medical Center, 99 TW Alexander Drive, P.O. Box 14991, Research Triangle Park, NC, 27709, USA
syoun@duke.edu

SOURCE: Analytical Biochemistry, (May 15 2003) Vol. 316, No. 2, pp. 175-180. print.

ISSN: 0003-2697 (ISSN print).

DOCUMENT TYPE: Article

LANGUAGE: English

ENTRY DATE: Entered STN: 25 Jun 2003

Last Updated on STN: 1 Aug 2003

ABSTRACT: Patients with glycogen storage disease type II (GSD II) typically excrete increased amounts of a glycogen-derived glucose **tetrasaccharide**, Glc4alpha1-6Glc4alpha1-4Glc4alpha1-4Glc (Glc4), in the urine. With the advent of a new enzyme replacement therapy for GSD II, there is a need for early

identification of patients with this disease and for monitoring the efficacy of treatment. Glc4 is a good candidate biomarker for GSD II. A simple and robust method using stable isotope dilution-electrospray ionization-tandem mass spectrometry for the analysis of Glc4 in biological samples was developed. A ¹³C6-labeled stable isotope internal standard was synthesized by transglycosylation using a recombinant alpha-amylase. Butyl 4-aminobenzoate derivatives of Glc4 and the internal standard were analyzed using multiple reaction monitoring. This method was shown to be accurate and precise by the repeated analysis of calibrators and quality control samples in urine and plasma. There was good agreement with a high-performance liquid chromatography-UV method for urine samples, whereas there was less agreement with plasma samples. Accurate determination from dried urine spot samples was also demonstrated. This method is amenable to high-throughput analysis, a necessary prerequisite for mass screening for GSD II.

CONCEPT CODE: Genetics - Human 03508
 Biochemistry studies - General 10060
 Biochemistry studies - Carbohydrates 10068
 Pathology - Therapy 12512
 Metabolism - General metabolism and metabolic pathways
 13002
 Metabolism - Metabolic disorders 13020
 Blood - Blood and lymph studies 15002
 Blood - Blood cell studies 15004
 Urinary system - Physiology and biochemistry 15504

INDEX TERMS: Major Concepts
 Biochemistry and Molecular Biophysics; Medical Genetics
 (Allied Medical Sciences); Metabolism; Methods and
 Techniques

INDEX TERMS: Parts, Structures, & Systems of Organisms
 plasma: blood and lymphatics; urine: excretory system

INDEX TERMS: Diseases
 Pompe disease: genetic disease, metabolic
 disease
 Glycogen Storage Disease Type II (MeSH)

INDEX TERMS: Diseases
 glycogen storage disease type II: genetic disease,
 metabolic disease, therapy, GSD II
 Glycogen Storage Disease Type II (MeSH)

INDEX TERMS: Chemicals & Biochemicals
 butyl 4-aminobenzoate derivatives; glucose; glucose
 tetrasaccharide: analysis; recombinant
 alpha-amylase

INDEX TERMS: Methods & Equipment
 enzyme replacement therapy: clinical techniques,
 therapeutic and prophylactic techniques; high
 performance liquid chromatography-UV method:
 chromatographic techniques, laboratory techniques;
 high-throughput analysis: laboratory techniques;
 multiple reaction monitoring: laboratory techniques;
 stable isotope dilution-electrospray ionization tandem
 mass spectrometry: laboratory techniques, spectrum
 analysis techniques; transglycosylation: laboratory
 techniques

ORGANISM: Classifier
 Hominidae 86215
 Super Taxa
 Primates; Mammalia; Vertebrata; Chordata; Animalia
 Organism Name
 human (common): patient
 Taxa Notes
 Animals, Chordates, Humans, Mammals, Primates,
 Vertebrates

REGISTRY NUMBER: 94-25-7D (butyl 4-aminobenzoate derivatives)
 50-99-7Q (glucose)
 58367-01-4Q (glucose)

L3 ANSWER 2 OF 5 BIOSIS COPYRIGHT 2004 BIOLOGICAL ABSTRACTS INC. on STN
 ACCESSION NUMBER: 2002:178595 BIOSIS
 DOCUMENT NUMBER: PREV200200178595
 TITLE: Determination of oligosaccharides in **Pompe** disease by electrospray ionization tandem mass spectrometry.
 AUTHOR(S): Rozaklis, Tina; Ramsay, Steven L.; Whitfield, Phillip D.; Ranieri, Enzo; Hopwood, John J.; Meikle, Peter J. [Reprint author]
 CORPORATE SOURCE: Lysosomal Diseases Research Unit, Department of Chemical Pathology, Women's and Children's Hospital, 72 King William Road, North Adelaide, South Australia, 5006, Australia peter.meikle@adelaide.edu.au
 SOURCE: Clinical Chemistry, (January, 2002) Vol. 48, No. 1, pp. 131-139. print.
 CODEN: CLCHAU. ISSN: 0009-9147.
 DOCUMENT TYPE: Article
 LANGUAGE: English
 ENTRY DATE: Entered STN: 6 Mar 2002
 Last Updated on STN: 6 Mar 2002
 ABSTRACT: Background: The development of therapies for lysosomal storage disorders has created a need for biochemical markers to monitor the efficacy of therapy and methods to quantify these markers in biologic samples. In *****Pompe***** disease, the concentration of a **tetrasaccharide**, consisting of four glucose residues, is reputedly increased in urine and plasma, but faster and more sensitive methods are required for the analysis of this, and other oligosaccharides, from biologic fluids. Methods: We optimized the derivatization of storage oligosaccharides with 1-phenyl-3-methyl-5-pyrazolone for the measurement, by electrospray ionization tandem mass spectrometry, of oligosaccharide concentrations in urine (n = 6), plasma (n = 11), and dried-blood spots (n = 17) from **Pompe**-affected individuals. Age-matched control samples of urine (n = 10), plasma (n = 28), and blood spots (n = 369) were also analyzed. Results: The mean **tetrasaccharide** concentration was increased in urine from infantile-onset (0.69-12 mmol/mol of creatinine) and adult-onset (0.22-3.0 mmol/mol of creatinine) **Pompe** individuals compared with age-matched controls. In plasma samples, an increased **tetrasaccharide** concentration was observed in some infantile patients (up to 22 mmol/L) compared with age-matched controls (mean, 2.2 mmol/L). The method developed was sensitive enough to determine oligosaccharide concentrations in a single 3-mm blood spot, but no differences were observed between blood spots from control and **Pompe**-affected individuals. Conclusions: Measurements of oligosaccharide concentrations in urine by this new method have potential application for the diagnosis and monitoring of patients with **Pompe** disease. Plasma analysis may have limited application for infantile patients, but analysis of blood spots does not discriminate between controls and affected individuals.
 CONCEPT CODE: Genetics - Human 03508
 Clinical biochemistry - General methods and applications 10006
 Biochemistry studies - Proteins, peptides and amino acids 10064
 Biochemistry studies - Carbohydrates 10068
 Metabolism - Metabolic disorders 13020
 Blood - Blood and lymph studies 15002
 Blood - Blood cell studies 15004
 Urinary system - Physiology and biochemistry 15504
 Pediatrics - 25000
 INDEX TERMS: Major Concepts
 Clinical Chemistry (Allied Medical Sciences); Medical Genetics (Allied Medical Sciences); Methods and Techniques
 INDEX TERMS: Parts, Structures, & Systems of Organisms
 blood: blood and lymphatics; plasma: blood and lymphatics; urine: excretory system

INDEX TERMS: Diseases
 Pompe disease: genetic disease, metabolic disease
 Glycogen Storage Disease Type II (MeSH)

INDEX TERMS: Diseases
 lysosomal storage disorder: metabolic disease

INDEX TERMS: Chemicals & Biochemicals
 1-phenyl-3-methyl-5-pyrazolone; creatinine;
 oligosaccharides: determination; **tetrasaccharide**

INDEX TERMS: Methods & Equipment
 electrospray ionization tandem mass spectrometry:
 determination method; plasma analysis: analytical method

ORGANISM: Classifier
 Hominidae 86215
 Super Taxa
 Primates; Mammalia; Vertebrata; Chordata; Animalia
 Organism Name
 human: adult, child, infant, patient
 Taxa Notes
 Animals, Chordates, Humans, Mammals, Primates, Vertebrates

REGISTRY NUMBER: 89-25-8Q (1-phenyl-3-methyl-5-pyrazolone)
 19735-89-8Q (1-phenyl-3-methyl-5-pyrazolone)
 60-27-5 (creatinine)

L3 ANSWER 3 OF 5 BIOSIS COPYRIGHT 2004 BIOLOGICAL ABSTRACTS INC. on STN
 ACCESSION NUMBER: 2001:554213 BIOSIS
 DOCUMENT NUMBER: PREV200100554213
 TITLE: Validation of glucose **tetrasaccharide** as a
 biomarker for diagnosis and monitoring enzyme replacement
 therapy for **Pompe** disease.

AUTHOR(S): An, Y. [Reprint author]; Millington, D. S. [Reprint
 author]; Kishnani, P. [Reprint author]; Amalfitano, A.
 [Reprint author]; Chen, Y. T. [Reprint author]

CORPORATE SOURCE: Division of Medical Genetics, Department of Pediatrics,
 Duke Univ Medical Center, Durham, NC, USA

SOURCE: American Journal of Human Genetics, (October, 2001) Vol.
 69, No. 4 Supplement, pp. 482. print.
 Meeting Info.: 51st Annual Meeting of the American Society
 of Human Genetics. San Diego, California, USA. October
 12-16, 2001.
 CODEN: AJHGAG. ISSN: 0002-9297.

DOCUMENT TYPE: Conference; (Meeting)
 Conference; Abstract; (Meeting Abstract)
 Conference; (Meeting Poster)

LANGUAGE: English

ENTRY DATE: Entered STN: 28 Nov 2001
 Last Updated on STN: 25 Feb 2002

CONCEPT CODE: General biology - Symposia, transactions and proceedings
 00520
 Genetics - General 03502
 Genetics - Animal 03506
 Pathology - Therapy 12512
 Metabolism - Metabolic disorders 13020
 Blood - Blood and lymph studies 15002
 Blood - Blood cell studies 15004
 Urinary system - Physiology and biochemistry 15504

INDEX TERMS: Major Concepts
 Genetics

INDEX TERMS: Parts, Structures, & Systems of Organisms
 blood: blood and lymphatics; urine: excretory system

INDEX TERMS: Diseases
 Pompe disease: genetic disease, metabolic disease
 Glycogen Storage Disease Type II (MeSH)

INDEX TERMS: Chemicals & Biochemicals
 acid a-glucosidase; glucose **tetrasaccharide**:
 biomarker
 INDEX TERMS: Methods & Equipment
 enzyme replacement therapy: therapeutic method
 INDEX TERMS: Miscellaneous Descriptors
 Meeting Abstract; Meeting Poster
 ORGANISM: Classifier
 Galliformes 85536
 Super Taxa
 Aves; Vertebrata; Chordata; Animalia
 Organism Name
 quail: animal model
 Taxa Notes
 Animals, Birds, Chordates, Nonhuman Vertebrates,
 Vertebrates

L3 ANSWER 4 OF 5 BIOSIS COPYRIGHT 2004 BIOLOGICAL ABSTRACTS INC. on STN
 ACCESSION NUMBER: 2001:26713 BIOSIS
 DOCUMENT NUMBER: PREV200100026713
 TITLE: Liquid chromatographic assay for a glucose
 tetrasaccharide, a putative biomarker for the
 diagnosis of **Pompe** disease.
 AUTHOR(S): An, Yan; Young, Sarah P.; Hillman, Stephen L.; Van Hove,
 Johan L. K. [Reprint author]; Chen, Yuan-Tsong; Millington,
 David S.
 CORPORATE SOURCE: Division of Medical Genetics, Department of Pediatrics,
 Duke University Medical Center, 99 TW Alexander Drive,
 Research Triangle Park, NC, 27709, USA
 vanan@acpub.duke.edu
 SOURCE: Analytical Biochemistry, (December 1, 2000) Vol. 287, No.
 1, pp. 136-143. print.
 CODEN: ANBCA2. ISSN: 0003-2697.
 DOCUMENT TYPE: Article
 LANGUAGE: English
 ENTRY DATE: Entered STN: 10 Jan 2001
 Last Updated on STN: 12 Feb 2002

ABSTRACT: A HPLC method associated with butyl-p-aminobenzoate derivatization has
 been developed for the analysis of a tetraglucose oligomer,
 Glc α 1-6Glc α 1-4Glc α 1-4Glc, designated Glc4, in biological fluids.
 This tetraglucose, normally excreted in the urine, has previously been shown to
 be elevated in a number of pathological conditions including **Pompe**
 disease (glycogen storage disease type II), which is caused by a deficiency of
 the lysosomal enzyme acid alpha-glucosidase. Concentrations of Glc4 in both
 urine and plasma were established for the age ranges of <1, 1-5, 6-10, 11-20,
 and >20 years, both in normal individuals and in a cohort of 21 patients with
 enzymatically confirmed **Pompe** disease. The Glc4 concentration
 decreased with age in both groups, but all the patients had elevated Glc4
 levels compared with age-matched controls. Electrospray tandem mass
 spectrometry was employed to establish the homogeneity of the HPLC peak for
 Glc4 and to investigate the identity of other unusual oligosaccharides excreted
 in patient urine. Our results demonstrate that this method is suitable for
 application in clinical laboratories to help establish the diagnosis of
 *****Pompe***** disease.

CONCEPT CODE: Pathology - Diagnostic 12504
 Genetics - Human 03508
 Clinical biochemistry - General methods and applications
 10006
 Metabolism - Metabolic disorders 13020

INDEX TERMS: Major Concepts
 Clinical Chemistry (Allied Medical Sciences); Methods
 and Techniques

INDEX TERMS: Diseases
 Pompe disease: genetic disease, metabolic
 disease

INDEX TERMS: Glycogen Storage Disease Type II (MeSH)
 Chemicals & Biochemicals
 glucose **tetrasaccharide**: assay, biomarker

INDEX TERMS: Methods & Equipment
 electrospray tandem mass spectrometry: analytical
 method, mass spectrometry: CB; high performance liquid
 chromatography [HPLC]: diagnostic method, liquid
 chromatography

ORGANISM: Classifier
 Hominidae 86215
 Super Taxa
 Primates; Mammalia; Vertebrata; Chordata; Animalia
 Organism Name
 human
 Taxa Notes
 Animals, Chordates, Humans, Mammals, Primates,
 Vertebrates

L3 ANSWER 5 OF 5 BIOSIS COPYRIGHT 2004 BIOLOGICAL ABSTRACTS INC. on STN
 ACCESSION NUMBER: 1991:6034 BIOSIS
 DOCUMENT NUMBER: PREV199191006034; BA91:6034
 TITLE: LEUKOCYTE ALPHA-1 4 GLUCOSIDASE AND ALPHA-1 6 GLUCOSIDASE
 ACTIVITIES TOWARDS OLIGOSACCHARIDES IN LATE ONSET
 GLYCOGENOSIS TYPE II.
 AUTHOR(S): KURIYAMA M [Reprint author]; HIWATARI R-I; OSAME M; IGATA A
 CORPORATE SOURCE: THIRD DEP INTERN MED, KAGOSHIMA UNIV SCH MED, KAGOSHIMA
 890, JPN
 SOURCE: Tohoku Journal of Experimental Medicine, (1990) Vol. 161,
 No. 4, pp. 343-351.
 CODEN: TJEMAO. ISSN: 0040-8727.
 DOCUMENT TYPE: Article
 FILE SEGMENT: BA
 LANGUAGE: ENGLISH
 ENTRY DATE: Entered STN: 8 Dec 1990
 Last Updated on STN: 30 Jan 1991

ABSTRACT: We describe the partial characterization and some properties of
 leucocyte α -glucosidase towards disaccharides with the α -1, 4
 (maltose) and α -1, 6-glucosidic linkage (isomaltose) and
 tetrasaccharides with the α -1, 4 (maltotetraose) and
 α -1, 6-i glucosidic linkage (**tetrasaccharide**, Glcal
 \rightarrow 6Glcal \rightarrow 4Glcal \rightarrow 4Glc, which was isolated
 from the urine of a patient with glycogenosis type II). Leucocyte
 α -glucosidase showed optimal activity towards all four oligosaccharides
 under two conditions, acidic (pH 4.0-4.5) and neutral (pH 6.0-6.5) regions.
 Our comparative studies on enzyme kinetics showed that leucocyte
 α -glucosidase was able to hydrolyze both the 1 \rightarrow 4 isomers and the
 1 \rightarrow 6 isomers at acidic and neutral pH. Acid α -glucosidase could
 hydrolyze maltose about 10 times faster than isomaltose, and maltotetraose
 about 5 times faster than **tetrasaccharide** isolated from urine. In
 leucocytes of the patient with late onset glycogenosis type II, acid
 α -glucosidase activities towards maltose, isomaltose, maltotetraose and
 tetrasaccharide isolated from urine showed 75.3%, 67.4%, 76.5% and
 41.4% of normal control values, respectively. Neutral α -glucosidase
 activities towards these four oligosaccharides were normal.
 Tetrasaccharide with α -1, 6-glucosidic linkage might be
 accumulated by the impaired hydrolysis in the circulation as well as the
 leakage of undegraded glycogen to the circulation from the affected muscle.

CONCEPT CODE: Genetics - Human 03508
 Clinical biochemistry - General methods and applications
 10006
 Biochemistry studies - Carbohydrates 10068
 Metabolism - Carbohydrates 13004
 Metabolism - Metabolic disorders 13020
 Blood - Blood cell studies 15004
 Blood - Lymphatic tissue and reticuloendothelial system

15008
Muscle - Pathology 17506

INDEX TERMS: Major Concepts
Blood and Lymphatics (Transport and Circulation);
Clinical Chemistry (Allied Medical Sciences);
Metabolism; Muscular System (Movement and Support)

INDEX TERMS: Miscellaneous Descriptors
HUMAN MUSCLE POMPE'S DISEASE

ORGANISM: Classifier
Hominidae 86215
Super Taxa
Primates; Mammalia; Vertebrata; Chordata; Animalia
Taxa Notes
Animals, Chordates, Humans, Mammals, Primates,
Vertebrates

REGISTRY NUMBER: 9001-42-7 (ALPHA-1,4-GLUCOSIDASE)
37288-48-5 (ALPHA-1,6-GLUCOSIDASE)

=> file caplus

COST IN U.S. DOLLARS	SINCE FILE ENTRY	TOTAL SESSION
FULL ESTIMATED COST	27.10	27.31

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FILE COVERS 1907 - 10 Mar 2004 VOL 140 ISS 11
FILE LAST UPDATED: 9 Mar 2004 (20040309/ED)

This file contains CAS Registry Numbers for easy and accurate substance identification.

=> s pompe

248 POMPE
15 POMPES
L4 250 POMPE
(POMPE OR POMPES)

=> s l4 and (tetrasaccharide)

2456 TETRASACCHARIDE
850 TETRASACCHARIDES
2996 TETRASACCHARIDE
(TETRASACCHARIDE OR TETRASACCHARIDES)
L5 8 L4 AND (TETRASACCHARIDE)

=> d l5 kwic 1-8

L5 ANSWER 1 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN
TI Analysis of a glucose **tetrasaccharide** elevated in **Pompe** disease by stable isotope dilution-electrospray ionization tandem mass

spectrometry

AB Patients with glycogen storage disease type II (GSD II) typically excrete increased amts. of a glycogen-derived glucose **tetrasaccharide**, Glc α 1-6Glc α 1-4Glc α 1-4Glc (Glc4), in the urine. With the advent of a new enzyme replacement therapy for GSD II, there is a . . .

ST glucose **tetrasaccharide** mass spectrometry glycogen storage disease urine

IT Blood analysis
Blood plasma
HPLC
Human
Isotope dilution mass spectrometry
Quality control
Urine analysis
(anal. of a glucose **tetrasaccharide** elevated in **Pompe** disease by stable isotope dilution-electrospray ionization tandem mass spectrometry)

IT Tandem mass spectrometry
(electrospray-ionization; anal. of a glucose **tetrasaccharide** elevated in **Pompe** disease by stable isotope dilution-electrospray ionization tandem mass spectrometry)

IT Electrospray ionization mass spectrometry
(tandem; anal. of a glucose **tetrasaccharide** elevated in **Pompe** disease by stable isotope dilution-electrospray ionization tandem mass spectrometry)

IT Glycogen storage disease
(type II; anal. of a glucose **tetrasaccharide** elevated in **Pompe** disease by stable isotope dilution-electrospray ionization tandem mass spectrometry)

IT 35175-16-7
RL: ANT (Analyte); DGN (Diagnostic use); ANST (Analytical study); BIOL (Biological study); USES (Uses)
(anal. of a glucose **tetrasaccharide** elevated in **Pompe** disease by stable isotope dilution-electrospray ionization tandem mass spectrometry)

L5 ANSWER 2 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

TI Determination of oligosaccharides in **pompe** disease by electrospray ionization tandem mass spectrometry

AB . . . need for biochem. markers to monitor the efficacy of therapy and methods to quantify these markers in biol. samples. In **Pompe** disease, the concentration of a **tetrasaccharide**, consisting of four glucose residues, is reputedly increased in urine and plasma, but faster and more sensitive methods are required. . . spectrometry, of oligosaccharide concns. in urine (n = 6), plasma (n = 11), and dried-blood spots (n = 17) from **Pompe**-affected individuals. Age-matched control samples of urine (n = 10), plasma (n = 28), and blood spots (n = 369) were also analyzed. Results: The mean **tetrasaccharide** concentration was increased in urine from infantile-onset (0.69-12 mmol/ mol. of creatinine) and adult-onset (0.22-3.0 mmol/mol of creatinine) **Pompe** individuals compared with age-matched controls. In plasma samples, an increased **tetrasaccharide** concentration was observed in some infantile patients (up to 22 μ mol/L) compared with age-matched controls (mean, 2.2 μ mol/L). The method. . . determine oligosaccharide concns. in a single 3-mm blood spot, but no differences were observed between blood spots from control and **Pompe**-affected individuals. Conclusions: Measurements of oligosaccharide concns. in urine by this new method have potential application for the diagnosis and monitoring of patients with **Pompe** disease. Plasma anal. may have limited application for infantile patients, but anal. of blood spots does not discriminate between controls. . .

ST oligosaccharide **pompe** disease electrospray ionization tandem mass spectrometry

IT Blood analysis

Blood plasma
 Body fluid
 Diagnosis
 Human
 Tandem mass spectrometry
 Urine analysis
 (oligosaccharides determination in **pompe** disease by electrospray ionization tandem mass spectrometry)

IT Oligosaccharides, analysis
 RL: ANT (Analyte); THU (Therapeutic use); ANST (Analytical study); BIOL (Biological study); USES (Uses)
 (oligosaccharides determination in **pompe** disease by electrospray ionization tandem mass spectrometry)

IT Glycogen storage disease
 (type II; oligosaccharides determination in **pompe** disease by electrospray ionization tandem mass spectrometry)

IT 89-25-8, 1-Phenyl-3-methyl-5-pyrazolone
 RL: BUU (Biological use, unclassified); BIOL (Biological study); USES (Uses)
 (oligosaccharides determination in **pompe** disease by electrospray ionization tandem mass spectrometry)

L5 ANSWER 3 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

TI Diagnostic methods for **Pompe** disease and other glycogen storage diseases

AB The invention concerns methods of screening subjects for lysosomal storage diseases, preferably glycogen storage diseases, using a **tetrasaccharide** as a biomarker. In a more preferred embodiment, subjects are screened for **Pompe** disease (i.e., glycogen storage disease type II). Also provided are neonatal screening assays. The present invention further provides methods of monitoring the clin. condition and efficacy of therapeutic treatment in affected subjects. Further provided are methods of measuring a **tetrasaccharide** biomarker by tandem mass spectrometry, preferably, as part of a neonatal screening assay for **Pompe** disease.

ST glycogen storage **Pompe** disease diagnosis **tetrasaccharide**
 HPLC mass spectrometry

IT Amniotic fluid
 Animal tissue
 Biomarkers (biological responses)
 Blood analysis
 Blood plasma
 Blood serum
 Body fluid
 Cell
 Electrospray ionization mass spectrometry
 Glycogen storage disease
 HPLC
 Human
 Immunoassay
 Liquid chromatography
 Lysosomal storage disease
 Mass spectrometry
 Newborn
 Purification
 Sputum
 Tandem mass spectrometry
 Urine analysis
 (diagnostic methods for **Pompe** disease and other glycogen storage diseases)

IT Blood analysis
 (dried blood spot; diagnostic methods for **Pompe** disease and other glycogen storage diseases)

IT Standard substances, analytical
 (internal; diagnostic methods for **Pompe** disease and other

glycogen storage diseases)

IT Ecology
(population; diagnostic methods for **Pompe** disease and other glycogen storage diseases)

IT Glycogen storage disease
(type II; diagnostic methods for **Pompe** disease and other glycogen storage diseases)

IT Glycogen storage disease
(type III; diagnostic methods for **Pompe** disease and other glycogen storage diseases)

IT Glycogen storage disease
(type VI; diagnostic methods for **Pompe** disease and other glycogen storage diseases)

IT 35175-16-7 379261-84-4
RL: ANT (Analyte); DGN (Diagnostic use); ANST (Analytical study); BIOL (Biological study); USES (Uses)
(diagnostic methods for **Pompe** disease and other glycogen storage diseases)

IT 89-25-8, 1-Phenyl-3-methyl-5-pyrazolone 93-97-0, Benzoic anhydride 94-25-7, Butyl p-aminobenzoate 27918-14-5, 2-Aminoacridone
RL: ARG (Analytical reagent use); ANST (Analytical study); USES (Uses)
(diagnostic methods for **Pompe** disease and other glycogen storage diseases)

IT 34620-76-3, Maltopentaose 34620-77-4, Maltohexaose 379261-83-3
RL: ARU (Analytical role, unclassified); ANST (Analytical study)
(diagnostic methods for **Pompe** disease and other glycogen storage diseases)

L5 ANSWER 4 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

TI Liquid chromatographic assay for a glucose **tetrasaccharide**, a putative biomarker for the diagnosis of **pompe** disease

AB . . . tetraglucose, normally excreted in the urine, has previously been shown to be elevated in a number of pathol. conditions including **Pompe** disease (glycogen storage disease type II), which is caused by a deficiency of the lysosomal enzyme acid α -glucosidase. Concns. of. . . 1-5, 6-10, 11-20, and >20 yr, both in normal individuals and in a cohort of 21 patients with enzymically confirmed **Pompe** disease. The Glc4 concentration decreased with age in both groups, but all the patients had elevated Glc4 levels compared with. . . urine. Our results demonstrate that this method is suitable for application in clin. labs. to help establish the diagnosis of **Pompe** disease. (c)
2000 Academic Press.

ST glucose **tetrasaccharide** detn HPLC **Pompe** disease

IT HPLC
Urine analysis
(glucose **tetrasaccharide** detn by HPLC for diagnosis of **Pompe** disease)

IT Glycogen storage disease
(type II; glucose **tetrasaccharide** detn by HPLC for diagnosis of **Pompe** disease)

IT 35175-16-7
RL: ANT (Analyte); THU (Therapeutic use); ANST (Analytical study); BIOL (Biological study); USES (Uses)
(tetraglucose; glucose **tetrasaccharide** detn by HPLC for diagnosis of **Pompe** disease)

L5 ANSWER 5 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

AB . . . the partial characterization and some properties of leukocyte α -glucosidase towards disaccharides with the α -1,4 (maltose) and α -1,6-glucosidic linkage (isomaltose) and **tetrasaccharides** with the α -1,4 (maltotetraose) and α -1,6-glucosidic linkage (**tetrasaccharide**, Glc α 1 \rightarrow 6Glc α 1 \rightarrow 4Glc α 1 \rightarrow 4Glc, which was isolated from the urine of a patient with glycogenosis type II). Leukocyte α -glucosidase showed optimal activity towards. . .

neutral pH. Acid α -glucosidase could hydrolyze maltose about 10 times faster than isomaltose, and maltotetraose about 5 times faster than **tetrasaccharide** isolated from urine. In leukocytes of the patient with late onset glycogenosis type II, acid α -glucosidase activities towards maltose, isomaltose, maltotetraose and **tetrasaccharide** isolated from urine showed 75.3%, 67.4%, 76.5% and 41.4% of normal control values, resp. Neutral α -glucosidase activities towards these four oligosaccharides were normal. **Tetrasaccharide** with α -1,6-glucosidic linkage might be accumulated by the impaired hydrolysis in the circulation as well as the leakage of undergraded. . .

IT Leukocyte

(α -glucosidase of, in **Pompe's** disease in human, oligosaccharides hydrolysis by)

IT Glycogenosis

(**Pompe's** disease, α -glucosidase of leukocytes in, in human, oligosaccharides hydrolysis by)

L5 ANSWER 6 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

AB, are specifically bound by the antibody with similar affinities, but the affinity is somewhat higher for chains containing the **tetrasaccharide** sequence Glc α 1-6Glc α 1-4Glc α 1-4Glc at the nonreducing terminal. Utilization of affinity methods offers clear advantages for isolation and characterization of oligosaccharides with. . . .

IT Glycogenosis

(**Pompe's** disease, oligodextrins of urine in, structural and immunochem. anal. of)

L5 ANSWER 7 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

TI Thin-layer chromatography of oligosaccharides in urine as a rapid indication for the diagnosis of lysosomal acid maltase deficiency (**Pompe's** disease)

AB desalted by ion-exchange chromatog. and then analyzed by the TLC method of R. Humbel and M. Collart (1975). The glucose-containing **tetrasaccharide** α -D-Glc-(1 \rightarrow 6)- α -D-Glc-(1 \rightarrow 4)- α -D-Glc-(1 \rightarrow 4)-D-Glc was detected in urine of patients with **Pompe's** disease. This method is potentially useful for the rapid and reliable diagnosis of lysosomal acid maltase deficiency. However, lactobionic acid. . . .

ST urine oligosaccharide thin layer chromatog; **Pompe** disease diagnosis oligosaccharide TLC; lysosomal acid maltase deficiency diagnosis; desalting urine oligosaccharide TLC

IT Urine analysis

(oligosaccharides detection in, of humans by TLC in **Pompe's** disease diagnosis, desalting in)

IT Glycogenosis

(**Pompe's** disease, diagnosis of, oligosaccharides of human urine in)

IT 35175-16-7

RL: ANT (Analyte); ANST (Analytical study)

(detection of, in human urine by TLC in **Pompe's** disease diagnosis)

IT 96-82-2

RL: ANST (Analytical study)

(interference by, in oligosaccharides detection in human urine by TLC in **Pompe's** disease diagnosis)

L5 ANSWER 8 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

TI Increased excretion of a glucose-containing **tetrasaccharide** in the urine of a patient with glycogen storage disease type II (**Pompe's** disease)

AB A **tetrasaccharide** was isolated from the urine (13 mg/24 hr) of a 10-year-old boy with the childhood form of the title disease.. . . . amts. from normal urine. Larger glucose-containing oligosaccharides, not detected in normal urine, were also present in the urine of the

Pompe case.
ST **Pompe** disease carbohydrate structure; glycogen storage disease
tetrasaccharide urine; urine glycogen storage disease
IT Glycogenosis
(glucose-containing **tetrasaccharide** of urine in)

=> d 15 iall 1-8

L5 ANSWER 1 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN
ACCESSION NUMBER: 2003:311407 CAPLUS
DOCUMENT NUMBER: 140:90078
ENTRY DATE: Entered STN: 23 Apr 2003
TITLE: Analysis of a glucose **tetrasaccharide**
elevated in **Pompe** disease by stable isotope
dilution-electrospray ionization tandem mass
spectrometry
AUTHOR(S): Young, Sarah P.; Stevens, Robert D.; An, Yan; Chen,
Yuan-Tsong; Millington, David S.
CORPORATE SOURCE: Department of Pediatrics, Division of Medical
Genetics, Biochemical Genetics Laboratory, Duke
University Medical Center, Research Triangle Park, NC,
27709, USA
SOURCE: Analytical Biochemistry (2003), 316(2), 175-180
CODEN: ANBCA2; ISSN: 0003-2697
PUBLISHER: Elsevier Science
DOCUMENT TYPE: Journal
LANGUAGE: English
CLASSIFICATION: 9-5 (Biochemical Methods)
Section cross-reference(s): 1

ABSTRACT:
Patients with glycogen storage disease type II (GSD II) typically excrete increased amts. of a glycogen-derived glucose **tetrasaccharide**, Glc α 1-6Glc α 1-4Glc α 1-4Glc (Glc4), in the urine. With the advent of a new enzyme replacement therapy for GSD II, there is a need for early identification of patients with this disease and for monitoring the efficacy of treatment. Glc4 is a good candidate biomarker for GSD II. A simple and robust method using stable isotope dilution-electrospray ionization-tandem mass spectrometry for the anal. of Glc4 in biol. samples was developed. A ¹³C6-labeled stable isotope internal standard was synthesized by transglycosylation using a recombinant α -amylase. Bu 4-aminobenzoate derivs. of Glc4 and the internal standard were analyzed using multiple reaction monitoring. This method was shown to be accurate and precise by the repeated anal. of calibrators and quality control samples in urine and plasma. There was good agreement with a high-performance liquid chromatog.-UV method for urine samples, whereas there was less agreement with plasma samples. Accurate determination from dried urine spot samples was also demonstrated. This method is amenable to high-throughput anal., a necessary prerequisite for mass screening for GSD II.

SUPPL. TERM: glucose **tetrasaccharide** mass spectrometry glycogen
storage disease urine
INDEX TERM: Blood analysis
Blood plasma
HPLC
Human
Isotope dilution mass spectrometry
Quality control
Urine analysis
(anal. of a glucose **tetrasaccharide** elevated in
Pompe disease by stable isotope
dilution-electrospray ionization tandem mass spectrometry)
INDEX TERM: Tandem mass spectrometry
(electrospray-ionization; anal. of a glucose

tetrasaccharide elevated in **Pompe** disease by stable isotope dilution-electrospray ionization tandem mass spectrometry)

INDEX TERM: Electrospray ionization mass spectrometry (tandem; anal. of a glucose **tetrasaccharide** elevated in **Pompe** disease by stable isotope dilution-electrospray ionization tandem mass spectrometry)

INDEX TERM: Glycogen storage disease (type II; anal. of a glucose **tetrasaccharide** elevated in **Pompe** disease by stable isotope dilution-electrospray ionization tandem mass spectrometry)

INDEX TERM: 35175-16-7
 ROLE: ANT (Analyte); DGN (Diagnostic use); ANST (Analytical study); BIOL (Biological study); USES (Uses)
 (anal. of a glucose **tetrasaccharide** elevated in **Pompe** disease by stable isotope dilution-electrospray ionization tandem mass spectrometry)

REFERENCE COUNT: 18 THERE ARE 18 CITED REFERENCES AVAILABLE FOR THIS RECORD.

REFERENCE(S): (1) Amalfitano, A; Genet Med 2001, V3, P132 CAPLUS
 (2) An, Y; Anal Biochem 2000, V287, P136 CAPLUS
 (3) Bland, J; Lancet 1986, V1(8476), P307 MEDLINE
 (4) De Praeter, C; Am J Hum Genet 2000, V66, P1744 CAPLUS
 (5) Hallgren, P; Eur J Clin Invest 1974, V4, P429 CAPLUS
 (6) Hallgren, P; J Biol Chem 1977, V252, P1034 CAPLUS
 (7) Kumlien, J; Clin Chim Acta 1988, V176, P39 CAPLUS
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 (9) Lennartson, G; Biomed Mass Spectrom 1976, V3, P51 CAPLUS
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 (12) Rozaklis, T; Clin Chem 2002, V48, P131 CAPLUS
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L5 ANSWER 2 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

ACCESSION NUMBER: 2002:15925 CAPLUS

DOCUMENT NUMBER: 136:131060

ENTRY DATE: Entered STN: 08 Jan 2002

TITLE: Determination of oligosaccharides in **pompe** disease by electrospray ionization tandem mass spectrometry

AUTHOR(S): Rozaklis, Tina; Ramsay, Steven L.; Whitfield, Phillip D.; Ranieri, Enzo; Hopwood, John J.; Meikle, Peter J.

CORPORATE SOURCE: Lysosomal Diseases Research Unit, Women's and Children's Hospital, North Adelaide, South Australia, 5006, Australia

SOURCE: Clinical Chemistry (Washington, DC, United States) (2002), 48(1), 131-139
 CODEN: CLCHAU; ISSN: 0009-9147

PUBLISHER: American Association for Clinical Chemistry

DOCUMENT TYPE: Journal

LANGUAGE: English

CLASSIFICATION: 9-5 (Biochemical Methods)
 Section cross-reference(s): 14

ABSTRACT:

Background: The development of therapies for lysosomal storage disorders has created a need for biochem. markers to monitor the efficacy of therapy and methods to quantify these markers in biol. samples. In **Pompe** disease, the concentration of a **tetrasaccharide**, consisting of four glucose residues, is reputedly increased in urine and plasma, but faster and more sensitive methods are required for the anal. of this, and other

oligosaccharides, from biol. fluids. Methods: We optimized the derivatization of storage oligosaccharides with 1-phenyl-3-methyl-5-pyrazolone for the measurement, by electrospray ionization tandem mass spectrometry, of oligosaccharide concns. in urine (n = 6), plasma (n = 11), and dried-blood spots (n = 17) from **Pompe**-affected individuals. Age-matched control samples of urine (n = 10), plasma (n = 28), and blood spots (n = 369) were also analyzed. Results: The mean **tetrasaccharide** concentration was increased in urine from infantile-onset (0.69-12 mmol/ mol. of creatinine) and adult-onset (0.22-3.0 mmol/mol of creatinine) **Pompe** individuals compared with age-matched controls. In plasma samples, an increased **tetrasaccharide** concentration was observed in some infantile patients (up to 22 μ mol/L) compared with age-matched controls (mean, 2.2 μ mol/L). The method developed was sensitive enough to determine oligosaccharide concns. in a single 3-mm blood spot, but no differences were observed between blood spots from control and **Pompe**-affected individuals. Conclusions: Measurements of oligosaccharide concns. in urine by this new method have potential application for the diagnosis and monitoring of patients with **Pompe** disease. Plasma anal. may have limited application for infantile patients, but anal. of blood spots does not discriminate between controls and affected individuals.

SUPPL. TERM: oligosaccharide **pompe** disease electrospray ionization tandem mass spectrometry

INDEX TERM: Blood analysis
Blood plasma
Body fluid
Diagnosis
Human
Tandem mass spectrometry
Urine analysis
(oligosaccharides determination in **pompe** disease by electrospray ionization tandem mass spectrometry)

INDEX TERM: Oligosaccharides, analysis
ROLE: ANT (Analyte); THU (Therapeutic use); ANST (Analytical study); BIOL (Biological study); USES (Uses)
(oligosaccharides determination in **pompe** disease by electrospray ionization tandem mass spectrometry)

INDEX TERM: Glycogen storage disease
(type II; oligosaccharides determination in **pompe** disease by electrospray ionization tandem mass spectrometry)

INDEX TERM: 89-25-8, 1-Phenyl-3-methyl-5-pyrazolone
ROLE: BUU (Biological use, unclassified); BIOL (Biological study); USES (Uses)
(oligosaccharides determination in **pompe** disease by electrospray ionization tandem mass spectrometry)

REFERENCE COUNT: 28 THERE ARE 28 CITED REFERENCES AVAILABLE FOR THIS RECORD.

REFERENCE(S): (1) An, Y; Anal Biochem 2000, V287, P136 CAPLUS
(2) Ausems, M; Eur J Hum Genet 1999, V7, P713 CAPLUS
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L5 ANSWER 3 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

ACCESSION NUMBER: 2001:904726 CAPLUS

DOCUMENT NUMBER: 136:17717

ENTRY DATE: Entered STN: 14 Dec 2001

TITLE: Diagnostic methods for **Pompe** disease and other glycogen storage diseases

INVENTOR(S): Millington, David S.; An, Yan; Chen, Yuan Tsong; Stevens, Robert D.; Young, Sarah P.; Van Hove, Johan L. K.

PATENT ASSIGNEE(S): Duke University, USA; Van Hove, Johan L. K.

SOURCE: PCT Int. Appl., 75 pp.

CODEN: PIXXD2

DOCUMENT TYPE: Patent

LANGUAGE: English

INT. PATENT CLASSIF.:

MAIN: G01N033-50

CLASSIFICATION: 9-16 (Biochemical Methods)

Section cross-reference(s): 14

FAMILY ACC. NUM. COUNT: 1

PATENT INFORMATION:

PATENT NO.	KIND	DATE	APPLICATION NO.	DATE
WO 2001094941	A2	20011213	WO 2001-US18288	20010606
WO 2001094941	A3	20030821		
W:	AE, AG, AL, AM, AT, AU, AZ, BA, BB, BG, BR, BY, BZ, CA, CH, CN, CO, CR, CU, CZ, DE, DK, DM, DZ, EC, EE, ES, FI, GB, GD, GE, GH, GM, HR, HU, ID, IL, IN, IS, JP, KE, KG, KP, KR, KZ, LC, LK, LR, LS, LT, LU, LV, MA, MD, MG, MK, MN, MW, MX, MZ, NO, NZ, PL, PT, RO, RU, SD, SE, SG, SI, SK, SL, TJ, TM, TR, TT, TZ, UA, UG, US, UZ, VN, YU, ZA, ZW, AM, AZ, BY, KG, KZ, MD, RU, TJ, TM			
RW:	GH, GM, KE, LS, MW, MZ, SD, SL, SZ, TZ, UG, ZW, AT, BE, CH, CY, DE, DK, ES, FI, FR, GB, GR, IE, IT, LU, MC, NL, PT, SE, TR, BF, BJ, CF, CG, CI, CM, GA, GN, GW, ML, MR, NE, SN, TD, TG			
US 2002102737	A1	20020801	US 2001-875327	20010606
EP 1360485	A2	20031112	EP 2001-944308	20010606
R:	AT, BE, CH, DE, DK, ES, FR, GB, GR, IT, LI, LU, NL, SE, MC, PT, IE, SI, LT, LV, FI, RO, MK, CY, AL, TR			
JP 2004501365	T2	20040115	JP 2002-502439	20010606
PRIORITY APPLN. INFO.:			US 2000-209920P P	20000607
			WO 2001-US18288 W	20010606

ABSTRACT:

The invention concerns methods of screening subjects for lysosomal storage diseases, preferably glycogen storage diseases, using a **tetrasaccharide** as a biomarker. In a more preferred embodiment, subjects are screened for *****Pompe***** disease (i.e., glycogen storage disease type II). Also provided are neonatal screening assays. The present invention further provides methods of monitoring the clin. condition and efficacy of therapeutic treatment in affected subjects. Further provided are methods of measuring a *****tetrasaccharide***** biomarker by tandem mass spectrometry, preferably, as part of a neonatal screening assay for **Pompe** disease.

Applicant

SUPPL. TERM: glycogen storage **Pompe** disease diagnosis
tetrasaccharide HPLC mass spectrometry

INDEX TERM: Amniotic fluid
 Animal tissue
 Biomarkers (biological responses)
 Blood analysis
 Blood plasma
 Blood serum
 Body fluid
 Cell
 Electrospray ionization mass spectrometry
 Glycogen storage disease
 HPLC
 Human
 Immunoassay
 Liquid chromatography
 Lysosomal storage disease
 Mass spectrometry
 Newborn
 Purification
 Sputum
 Tandem mass spectrometry
 Urine analysis
 (diagnostic methods for **Pompe** disease and other
 glycogen storage diseases)

INDEX TERM: Blood analysis
 (dried blood spot; diagnostic methods for **Pompe**
 disease and other glycogen storage diseases)

INDEX TERM: Standard substances, analytical
 (internal; diagnostic methods for **Pompe** disease
 and other glycogen storage diseases)

INDEX TERM: Ecology
 (population; diagnostic methods for **Pompe**
 disease and other glycogen storage diseases)

INDEX TERM: Glycogen storage disease
 (type II; diagnostic methods for **Pompe** disease
 and other glycogen storage diseases)

INDEX TERM: Glycogen storage disease
 (type III; diagnostic methods for **Pompe** disease
 and other glycogen storage diseases)

INDEX TERM: Glycogen storage disease
 (type VI; diagnostic methods for **Pompe** disease
 and other glycogen storage diseases)

INDEX TERM: 35175-16-7 379261-84-4
 ROLE: ANT (Analyte); DGN (Diagnostic use); ANST (Analytical
 study); BIOL (Biological study); USES (Uses)
 (diagnostic methods for **Pompe** disease and other
 glycogen storage diseases)

INDEX TERM: 89-25-8, 1-Phenyl-3-methyl-5-pyrazolone 93-97-0, Benzoic
 anhydride 94-25-7, Butyl p-aminobenzoate 27918-14-5,
 2-Aminoacridone
 ROLE: ARG (Analytical reagent use); ANST (Analytical study);
 USES (Uses)
 (diagnostic methods for **Pompe** disease and other
 glycogen storage diseases)

INDEX TERM: 34620-76-3, Maltopentaose 34620-77-4, Maltohexaose
 379261-83-3
 ROLE: ARU (Analytical role, unclassified); ANST (Analytical
 study)
 (diagnostic methods for **Pompe** disease and other
 glycogen storage diseases)

DOCUMENT NUMBER: 134:175062
ENTRY DATE: Entered STN: 15 Nov 2000
TITLE: Liquid chromatographic assay for a glucose
tetrasaccharide, a putative biomarker for the
diagnosis of **pompe** disease
AUTHOR(S): An, Yan; Young, Sarah P.; Hillman, Stéphen L.; Van
Hove, Johan L. K.; Chen, Yuan-Tsong; Millington, David
S.
CORPORATE SOURCE: Division of Medical Genetics, Department of
Pediatrics, Duke University Medical Center, Research
Triangle Park, NC, 27709, USA
SOURCE: Analytical Biochemistry (2000), 287(1), 136-143
CODEN: ANBCA2; ISSN: 0003-2697
PUBLISHER: Academic Press
DOCUMENT TYPE: Journal
LANGUAGE: English
CLASSIFICATION: 9-3 (Biochemical Methods)
Section cross-reference(s): 14

ABSTRACT:

A HPLC method associated with butyl-p-aminobenzoate derivatization has been developed for the anal. of a tetraglucose oligomer, Glc α 1-6Glc α 1-4Glc α 1-4Glc, designated Glc4, in biol. fluids. This tetraglucose, normally excreted in the urine, has previously been shown to be elevated in a number of pathol. conditions including **Pompe** disease (glycogen storage disease type II), which is caused by a deficiency of the lysosomal enzyme acid α -glucosidase. Concns. of Glc4 in both urine and plasma were established for the age ranges of <1, 1-5, 6-10, 11-20, and >20 yr, both in normal individuals and in a cohort of 21 patients with enzymically confirmed *****Pompe***** disease. The Glc4 concentration decreased with age in both groups, but all the patients had elevated Glc4 levels compared with age-matched controls. Electrospray tandem mass spectrometry was employed to establish the homogeneity of the HPLC peak for Glc4 and to investigate the identity of other unusual oligosaccharides excreted in patient urine. Our results demonstrate that this method is suitable for application in clin. labs. to help establish the diagnosis of **Pompe** disease. (c) 2000 Academic Press.

SUPPL. TERM: glucose **tetrasaccharide** detn HPLC **Pompe**
disease
INDEX TERM: HPLC
Urine analysis
(glucose **tetrasaccharide** detn by HPLC for
diagnosis of **Pompe** disease)
INDEX TERM: Glycogen storage disease
(type II; glucose **tetrasaccharide** detn by HPLC
for diagnosis of **Pompe** disease)
INDEX TERM: 35175-16-7
ROLE: ANT (Analyte); THU (Therapeutic use); ANST (Analytical
study); BIOL (Biological study); USES (Uses)
(tetraglucose; glucose **tetrasaccharide** detn by
HPLC for diagnosis of **Pompe** disease)
REFERENCE COUNT: 26 THERE ARE 26 CITED REFERENCES AVAILABLE FOR THIS
RECORD.
REFERENCE(S): (1) Amalfitano, A; Proc Natl Acad Sci 1999, V96, P8861
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L5 ANSWER 5 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

ACCESSION NUMBER: 1991:59967 CAPLUS

DOCUMENT NUMBER: 114:59967

ENTRY DATE: Entered STN: 23 Feb 1991

TITLE: Leukocyte α -1,4- and α -1,6-glucosidase activities towards oligosaccharides in late onset glycogenosis type II

AUTHOR(S): Kuriyama, Masaru; Hiwatari, Ryoichi; Osame, Mitsuhiro; Igata, Akihiro

CORPORATE SOURCE: Sch. Med., Kagoshima Univ., Kagoshima, 890, Japan

SOURCE: Tohoku Journal of Experimental Medicine (1990), 161(4), 343-51

CODEN: TJEMAO; ISSN: 0040-8727

DOCUMENT TYPE: Journal

LANGUAGE: English

CLASSIFICATION: 14-14 (Mammalian Pathological Biochemistry)

ABSTRACT:

This study describes the partial characterization and some properties of leukocyte α -glucosidase towards disaccharides with the α -1,4 (maltose) and α -1,6-glucosidic linkage (isomaltose) and ***tetrasaccharides*** with the α -1,4 (maltotetraose) and α -1,6-glucosidic linkage (**tetrasaccharide**, Glc α 1 \rightarrow 6Glc α 1 \rightarrow 4Glc α 1 \rightarrow 4Glc, which was isolated from the urine of a patient with glycogenosis type II). Leukocyte α -glucosidase showed optimal activity towards all four oligosaccharides in acidic (pH 4.0-4.5) and neutral (pH 6.0-6.5) regions. Leukocyte α -glucosidase was able to hydrolyze both the 1 \rightarrow 4 isomers and the 1 \rightarrow 6 isomers at acidic and neutral pH. Acid α -glucosidase could hydrolyze maltose about 10 times faster than isomaltose, and maltotetraose about 5 times faster than **tetrasaccharide** isolated from urine. In leukocytes of the patient with late onset glycogenosis type II, acid α -glucosidase activities towards maltose, isomaltose, maltotetraose and ***tetrasaccharide*** isolated from urine showed 75.3%, 67.4%, 76.5% and 41.4% of normal control values, resp. Neutral α -glucosidase activities towards these four oligosaccharides were normal. **Tetrasaccharide** with α -1,6-glucosidic linkage might be accumulated by the impaired hydrolysis in the circulation as well as the leakage of undergraded glycogen to the circulation from the affected muscle.

SUPPL. TERM: glycosidase oligosaccharide hydrolysis glycogenosis type II

INDEX TERM: Oligosaccharides

ROLE: RCT (Reactant); RACT (Reactant or reagent)
(hydrolysis of, by α -glucosidase of leukocytes in glycogenosis type II in human)

INDEX TERM: Leukocyte

(α -glucosidase of, in **Pompe's** disease in human, oligosaccharides hydrolysis by)

INDEX TERM: Glycogenosis
(**Pompe's** disease, α -glucosidase of leukocytes in, in human, oligosaccharides hydrolysis by)

INDEX TERM: 9001-42-7
ROLE: BIOL (Biological study)
(acid and neutral, of leukocyte in glycogenosis type II in human, oligosaccharides hydrolysis by)

INDEX TERM: 69-79-4, Maltose 499-40-1, Isomaltose 34612-38-9, Maltotetraose 35175-16-7
ROLE: RCT (Reactant); RACT (Reactant or reagent)
(hydrolysis of, by α -glucosidase of leukocytes in glycogenosis type II in human)

L5 ANSWER 6 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN

ACCESSION NUMBER: 1989:171227 CAPLUS

DOCUMENT NUMBER: 110:171227

ENTRY DATE: Entered STN: 12 May 1989

TITLE: Structural and immunochemical analysis of three α -limit dextrin oligosaccharides

AUTHOR(S): Kumlien, Johan; Groenberg, Gunnar; Nilsson, Bo; Maansson, Olle; Zopf, David; Lundblad, Arne

CORPORATE SOURCE: Dep. Clin. Chem., Univ. Hosp., Lund, S-221 85, Swed.

SOURCE: Archives of Biochemistry and Biophysics (1989), 269(2), 678-89

CODEN: ABBIA4; ISSN: 0003-9861

DOCUMENT TYPE: Journal

LANGUAGE: English

CLASSIFICATION: 14-14 (Mammalian Pathological Biochemistry)
Section cross-reference(s): 33

ABSTRACT:

Complete structures are described for 3 urinary oligodextrins from one patient with type II and one patient with type III glycogen storage disease. GLC-MS, direct probe MS, and ¹H NMR demonstrate two heptasaccharides and one hexasaccharide containing only α 1-4 and α 1-6 linkages. The observation that all 3 oligosaccharides were present in urine of both patients and the occurrence of α 1-4 and α 1-6 linkages in characteristic sequences indicates that the oligodextrins are limit dextrins derived from α -amylolytic degradation of glycogen. The binding affinities of the oligodextrins for a monoclonal antibody (401/6) raised against Glc α 1-6Glc α 1-4Glc α 1-4Glc, were determined by frontal anal. The highest affinity was exhibited by Glc α 1-6Glc α 1-4Glc α 1-4Glc followed by the 2 heptasaccharides with the hexasaccharide. The results from quant. affinity measurements agree with results of structural anal. by phys. methods in that all oligodextrins containing the nonreducing terminal sequence, Glc α 1-6Glc α 1-4Glc ..., are specifically bound by the antibody with similar affinities, but the affinity is somewhat higher for chains containing the ***tetrasaccharide*** sequence Glc α 1-6Glc α 1-4Glc α 1-4Glc at the nonreducing terminal. Utilization of affinity methods offers clear advantages for isolation and characterization of oligosaccharides with very similar structures.

SUPPL. TERM: dextrin oligosaccharide urine glycogenosis II III

INDEX TERM: Urine
(dextrin oligosaccharide of, in glycogenosis type II and type III in human)

INDEX TERM: Oligosaccharides
ROLE: BIOL (Biological study)
(of α -limit dextrin, of urine in glycogenosis type II and III in human)

INDEX TERM: Glycogenosis
(Cori's disease, oligodextrins of urine in, structural and immunochem. anal. of)

INDEX TERM: Glycogenosis

(Pompe's disease, oligodextrins of urine in,
structural and immunochem. anal. of)
INDEX TERM: 9004-53-9, Dextrin 40878-60-2 40878-61-3 40879-32-1
ROLE: PROC (Process)
(of urine, in glycogenosis type II and III in humans,
structural and immunochem. anal. of)

L5 ANSWER 7 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN
ACCESSION NUMBER: 1983:609089 CAPLUS
DOCUMENT NUMBER: 99:209089
ENTRY DATE: Entered STN: 12 May 1984
TITLE: Thin-layer chromatography of oligosaccharides in urine
as a rapid indication for the diagnosis of lysosomal
acid maltase deficiency (Pompe's disease)
AUTHOR(S): Blom, W.; Luteyn, J. C.; Kelholt-Dijkman, H. H.;
Huijmans, J. G. M.; Loonen, M. C. B.
CORPORATE SOURCE: Dep. Pediatr., Univ. Hosp. Rotterdam, Rotterdam, Neth.
SOURCE: Clinica Chimica Acta (1983), 134(1-2), 221-7
CODEN: CCATAR; ISSN: 0009-8981
DOCUMENT TYPE: Journal
LANGUAGE: English
CLASSIFICATION: 9-3 (Biochemical Methods)
Section cross-reference(s): 14

ABSTRACT:
Urine samples were desalted by ion-exchange chromatog. and then analyzed by the
TLC method of R. Humbel and M. Collart (1975). The glucose-containing
tetrasaccharide α -D-Glc-(1 \rightarrow 6)- α -D-Glc-(1 \rightarrow 4)-
 α -D-Glc-(1 \rightarrow 4)-D-Glc was detected in urine of patients with
Pompe 's disease. This method is potentially useful for the rapid and
reliable diagnosis of lysosomal acid maltase deficiency. However, lactobionic
acid can interfere with the method.

SUPPL. TERM: urine oligosaccharide thin layer chromatog; Pompe
disease diagnosis oligosaccharide TLC; lysosomal acid
maltase deficiency diagnosis; desalting urine
oligosaccharide TLC
INDEX TERM: Chromatography, thin-layer
(of oligosaccharides)
INDEX TERM: Urine analysis
(oligosaccharides detection in, of humans by TLC in
Pompe's disease diagnosis, desalting in)
INDEX TERM: Glycogenosis
(Pompe's disease, diagnosis of,
oligosaccharides of human urine in)
INDEX TERM: 9001-42-7
ROLE: ANST (Analytical study)
(deficiency of, diagnosis of, oligosaccharides of human
urine in)
INDEX TERM: 35175-16-7
ROLE: ANT (Analyte); ANST (Analytical study)
(detection of, in human urine by TLC in Pompe's
disease diagnosis)
INDEX TERM: 96-82-2
ROLE: ANST (Analytical study)
(interference by, in oligosaccharides detection in human
urine by TLC in Pompe's disease diagnosis)

L5 ANSWER 8 OF 8 CAPLUS COPYRIGHT 2004 ACS on STN
ACCESSION NUMBER: 1975:55516 CAPLUS
DOCUMENT NUMBER: 82:55516
ENTRY DATE: Entered STN: 12 May 1984
TITLE: Increased excretion of a glucose-containing
tetrasaccharide in the urine of a patient with
glycogen storage disease type II (Pompe's
disease)

AUTHOR(S): Hallgren, P.; Hansson, G.; Henriksson, K. C.; Hager, A.; Lundblad, Arne; Svensson, S.
 CORPORATE SOURCE: Inst. Med. Chem., Univ. Uppsala, Uppsala, Swed.
 SOURCE: European Journal of Clinical Investigation (1974), 4(6), 429-33
 CODEN: EJCIB8; ISSN: 0014-2972
 DOCUMENT TYPE: Journal
 LANGUAGE: English
 CLASSIFICATION: 14-3 (Mammalian Pathological Biochemistry)
 Section cross-reference(s): 13

ABSTRACT:

A **tetrasaccharide** was isolated from the urine (13 mg/24 hr) of a 10-year-old boy with the childhood form of the title disease. The structure, established by sugar anal., methylation anal., optical rotation, and enzymic degradation, was α -D-glucopyranosyl-(1 \rightarrow 6)- α -D-glucopyranosyl-(1 \rightarrow 4)- α -D-glucopyranosyl-(1 \rightarrow 4)-D-glucose. The same compound was also isolated in small amts. from normal urine. Larger glucose-containing oligosaccharides, not detected in normal urine, were also present in the urine of the **Pompe** case.

SUPPL. TERM: **Pompe** disease carbohydrate structure; glycogen storage disease **tetrasaccharide** urine; urine glycogen storage disease
 INDEX TERM: Glycogenosis
 (glucose-containing **tetrasaccharide** of urine in)
 INDEX TERM: Oligosaccharides
 ROLE: BIOL (Biological study)
 (of urine in glycogen storage disease)
 INDEX TERM: 35175-16-7
 ROLE: BIOL (Biological study)
 (of urine in glucogen storage disease)
 INDEX TERM: 25191-16-6
 ROLE: BIOL (Biological study)
 (oligomeric, of urine in glucogen storage disease)

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